CASE REPORT

Cervical unicentric Castleman’s disease in children

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KEYWORDS
Castleman’s disease; Lymph node; Neck; Child

Summary
Introduction: Unicentric Castleman’s disease (CD) is a rare benign lymphoproliferative disorder. A cervical location has rarely been reported in the pediatric literature.

Case report: A 13-year-old boy presented with a mass in the right neck region of 3 months’ evolution. Clinical examination revealed a smooth mobile 5 cm node deep in the sternocleidomastoid muscle. The diagnosis of hyaline vascular type CD was confirmed by complete surgical excision and histopathologic examination. Postoperative course was simple, without recurrence over 1 year’s follow-up.

Discussion and conclusion: Cervically located CD often shows few symptoms, and is rarely suggested by a cervical mass found in children. Definitive diagnosis is histopathological. The hyaline vascular type of unicentric CD has a good prognosis after complete resection of the lesion.

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Castleman’s disease (CD) is a benign lymphoproliferative disorder of unknown etiology [1,2]. In children, lesions are often mediastinal or abdominal [1], and only rarely cervical [3]. We report the case of a boy presenting with cervical unicentric CD (UCD).

Case report

A 13-year-old boy was admitted to the pediatric department for exploration of a right cervical mass, which had appeared 3 months previously and progressively increased in volume without any associated symptoms.

Clinical examination found a right cervical mass of 5 cm on its long axis, located along the vascular axis. It was supple, painless and relatively immobile, with normal skin cover. Biological analysis found no inflammatory syndrome.

Cervical ultrasound scan found multiple low right jugular carotid adenopathies, the largest of which was regularly contoured, hypoechoic, homogeneous and hypervasculnized, measuring 6 × 1.5 cm. Cervical MRI found two lesions (Fig. 1). The first was an oval spindle-shaped mass in the right juxtaocular carotid space, showing in isosignal in T1-weighted sequence, hypersignal in T2-weighted sequence, and with heterogeneous gadolinium uptake; it measured 4 × 2 × 6.5 cm, and pushed the carotid artery and internal jugular vein inward and forward and the neighbouring muscles outward, without signs of invasion. The second was a low right jugular carotid lesion of centimetric size, showing the same characteristics on imaging.

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Exploratory fine-needle aspiration indicated adenopathy without signs of malignancy. Both lesions were entirely removed on surgery. Macroscopic examination of the fresh specimen found an encapsulated oval mass of 6.5 × 3.5 × 1.3 cm; cross-sectional examination found either a fleshy or a yellowish white aspect, with a 2.3 cm long oblong nodule of similar aspect attached.

Microscopic examination found a hyperplastic lymph-node, the architecture of which was partially destroyed by hyperplastic lymphoid follicles with atrophied, fibrous and richly vascularized germ centers surrounded by concentric layers of small mature lymphocytes. The inter-follicle tissue was rich in capillary-type vessels. The histologic aspect was compatible with hyaline vascular UCD (Fig. 2).

Postoperative course was in this case simple, with no recurrence or metastasis over 1 year’s follow-up.

Discussion

CD was revealed on exploration of an isolated cervical mass. This illustrates the difficulty of diagnosing cervical UCD, especially in children: it is not only rare but also often relatively asymptomatic [1].

CD was first described in 1956, as a thymoma-like isolated mediastinal lymph-node hyperplasia [1,4]. It is a benign lymphoid proliferation, which has been variously named giant lymphoid hyperplasia, angiomatous lymphoid hamartoma, angiofollicular lymphoid hyperplasia, or giant benign lymphoma [1,4,5].

There are two forms of CD: unicentric, involving a single lymph-node site, and multicentric, involving several [6]. Etiology is unknown [7], although two theories have been suggested: immunological or dysembryoplastic [2]. Some authors consider CD to be secondary to immune deregulation induced by chronic antigen stimulation, or to a maladapted immune response to unidentified immune agents [2,7]. Onset may be at any age, including childhood, peaking in young adulthood [2]. In children, location is thoracic in 33% of cases, abdominal in 30%, cervical in 23% and axillary in 7% [1]. There have been few reports of cervical UCD in children [1], with only 30 observations: two children aged less than 1 year, 10 between 1 and 10 years, and 18 between 10 and 18 years [1–3]; the present patient, aged 13 years, was thus in the most frequent age group.

Diagnosis is confirmed postoperatively on histology [1,6]. Histologically, there are two forms of UCD: hyaline vascular and plasmacytic [6]; the former is more frequent in adults (90% of unicentric cases), whereas in children both are equally frequent [6].

Imaging in UCD is non-specific [4]. MRI, however, does show lesions in hypointense signal on T1-weighted images and in hypersignal on T2 [2]; it indicates benignity and determines local extension [3].

Unicentric forms of CD can often be successfully managed by complete surgical resection [5], obtaining recurrence-free resolution in almost all histologically hyaline vascular cases [1]. Two cases of cervical UCD showed spontaneous resolution, for no obvious reason [3]. Chemotherapy or radiation therapy, with or without associated surgery, should be considered for multicentric CD [1]; prognosis here is generally poor, with a few rare reports of malignant transformation [1,3].

Conclusion

Childhood cervical UCD is a rare benign clinicohistopathologic entity. Definitive diagnosis is anatomopathological, differentiating between hyaline vascular and plasmacytic forms. Treatment is surgical, with excellent prognosis in the hyaline vascular form if resection is total. Prolonged follow-up is essential, to detect possible recurrence or rare cases of malignant transformation.
Disclosure of interest

The authors declare that they have no conflicts of interest concerning this article.

References